The optimal Fontan operation: Lateral tunnel or extracardiac conduit?

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The most widely adopted variations of the Fontan procedure are the extracardiac conduit (ECC) and the lateral tunnel (LT). Over the last decade, there seemed to have been a shift in our practice, with the majority of the Fontan operations being the ECC, on the premise of the simplicity of this procedure and the expectation that it can better prevent the late occurrence of arrhythmias. A recent article in the *Journal* by a team from Boston demonstrated that in their hands, the ECC was associated with worse early outcomes compared with the LT.¹ We reviewed the existing literature to identify whether one of these techniques is superior to the other.

Here we performed a search of the PubMed database to identify studies with early and late outcomes. Inclusion criteria included (1) >200 patients, (2) published from 2005 onward, and (3) >50% with LT and ECC. Fourteen articles were identified for early outcomes, and 11 were identified for late outcomes (Tables 1 and 2).

First reported by Puga and colleagues¹⁸ in 1987 to address the issue of atrial septation, the LT technique became a major modification to the Fontan operation following a series of detailed hydrodynamic studies by de Leval.¹⁹ His work demonstrated that chamber pulsation (as with right atrial contraction), sharp corners, and regions of expansion increase the degree of turbulence, energy loss, and effective stenosis in the chamber and/or downstream vessel/conduit. De Leval and colleagues¹⁹ confirmed the hydrodynamic benefits of the linearized inferior vena cava (IVC) flow to the pulmonary vasculature through a lateral tunnel.

Humes and colleagues²⁰ and Marcelletti and colleagues²¹ and later reported the successful use of an extracardiac conduit. Its hypothesized benefits were the avoidance of intra-atrial prosthetic material, reduced risk of pulmonary venous obstruction, and potential reduced risk of supraventricular arrhythmias associated with atrial incisions, suture

The two most widely used Fontan variations: lateral tunnel and extracardiac conduit.

CENTRAL MESSAGE

Despite decades of follow-up, the optimal Fontan operation remains elusive. Given the variability in published outcomes, a randomized control trial might help identify the optimal technique.

See Commentaries on pages 1835 and 1836.

lines, and gradual distension of the atrial portion of the Fontan pathway.

FONTAN CIRCULATION FLUID DYNAMICS STUDIES

Minimizing Fontan pathway energy loss is paramount for avoiding both early and late complications. In addition, surgeons need to keep in mind that they have to construct the geometry of the anastomoses in a way that allows hepatic blood flow to both lungs, because the lack of this flow has been associated with the development of pulmonary arteriovenous malformations.

The work by de Leval and later confirmed by Sharma and colleagues²² and Lardo and colleagues²³ demonstrated that a degree of caval offset and flaring of the anastomosis can significantly reduce flow collision, recirculation, and subsequently energy loss.²⁴ Tang and colleagues²⁵ demonstrated significant variability within both LT and ECC based on the anastomotic positions and angulations.²⁵

Lardo and colleagues,²³ in their comparison of intra-atrial tunnel and extracardiac conduit, showed significantly lower energy loss in the ECC model. They also demonstrated that a mismatch in IVC–Fontan conduit size results in more energy loss, with a 1:1 size ratio as the most efficient. This further supports de Leval's original report

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TABLE 1. Early outcomes of contemporary techniques

Authors	Dates	Number	Early mortality, %	Comparison	Comment
Weixler et al, 2020 ¹	2000-2017	Total: 801 LT: 638 ECC: 183	Overall: 1.0 LT: 0.5 ECC: 3.1	Propensity-score matched: age, fenestration, heterotaxy, ventricular dominance and concomitant procedures	$P < .01$ in mortality in favor of LT.
Mery et al, 2019 ²	1995-2016	Total: 610 LT: 147 ECC: 463	Overall: 0.5	Demographics unavailable for Fontan-type comparison. Fontan type not considered a risk factor.	No difference reported in mortality or morbidity.
Downing et al, 2017 ³	1992-2009	Total: 773 LT: 409 ECC: 364	Overall: 3.5 LT: NR ECC: NR	Demographics unavailable for Fontan-type comparison. Fontan type included in multivariable risk factor analysis.	No difference reported in mortality.
Ono et al, 2016 ⁴	1994-2015	Total: 434 LT: 50 ECC: 384	Overall: 2 LT: 6 ECC: 2	ECC cohort had more right ventricular dominance, HLHS, and heterotaxy, and fewer concomitant procedures and fenestrations at time of Fontan.	$P = .02$ in mortality in favor of ECC.
Nakano et al, 2015 ⁵	1994-2014	Total: 500 LT: 0 ECC: 500	Overall: 0.4 LT: N/A ECC: 0.4	Only ECC	Only looked at ECC.
Iyengar et al, 2014 ⁶	1975-2010	Total: 1071 LT: 296 ECC: 536	Overall: 3.5 (including AP Fontan) LT: 2.0 (6/269) ECC: 1.3 (7/536)	Multivariable models accounting for common risk factors performed (including ventricular dominance and heterotaxy) to compare LT and ECC patients.	No difference reported in mortality.
Rogers et al, 2012 ⁷	1992-2009	Total: 771 LT: 409 ECC: 362	Overall: 3.5 LT: NR ECC: NR	Demographics unavailable for Fontan-type comparison. Multivariable analysis performed including all preoperative risk factors.	$P = .196$; no difference in mortality. ECC associated with prolonged pleural drainage and hospital stay (due to longer support times).
Stewart et al, 2012 ⁸	2000-2009	Total: 2747 LT: 1017 ECC: 1730	Overall: 1.6 LT: 0.9 ECC: 2.1	Unadjusted and multivariable analyses (age, weight, and all preoperative risk factors) performed, including logistic and linear regressions	$P = .02$ in mortality in favor of LT.
Robbers-Visser et al, 2010 ⁹	1988-2008	Total: 209 LT: 102 ECC: 107	Overall: 4 LT: 3.9 ECC: 4.7	Preoperative risk factors accounted for in multivariable analysis. HLHS and tricuspid atresia more common in ECC group.	No difference reported in mortality or morbidity.
Brown et al, 2010 ¹⁰	1992-2008	Total: 280 LT: 220 ECC: 0	Overall: 0.5 LT: 0.5 ECC: N/A	Only LT	Only looked at LT.
Salazar et al, 2010 ¹¹	2002-2008	Total: 226 LT: 69 ECC: 157	Overall: 1.3 LT: 1.4 ECC: 1.3	Full demographics unavailable for Fontan-type comparison. Risk factors included in multivariable analysis. No difference in concomitant procedures at the time of Fontan.	No difference in mortality. LT associated with longer ICU stay ($P = .022$) and total hospital stay ($P = .017$).

(Continued)

TABLE 1. Continued

Authors	Dates	Number	Early mortality, %	Comparison	Comment
Tweddell et al, 2009 ¹²	1994-2007	Total: 256 LT: 81 ECC: 175	Overall: 2 LT: NR ECC: NR	Multivariable analysis performed including preoperative risk factors (excluding heterotaxy).	No difference reported in mortality or morbidity.
Kim et al, 2008 ¹³	1996-2006	Total: 200 LT: 0 ECC: 200	Overall: 3 LT: N/A ECC: 3	Only LT	Only looked at ECC.
Hirsch et al, 2008 ¹⁴	1992-2007	Total: 636 LT: 585 ECC: 51	Overall: 4 LT: NR ECC: NR	ECC was performed in patients with anatomy not amenable to LT technique. Group comparison is not accurate due to selection bias.	$P = .02$ in mortality in favor of LT. ECC associated with prolonged effusions ($P = .01$) and atrial arrhythmias ($P = .02$). Complicated by minimal fenestration use in ECC (35%) vs 100% in LT.

LT, Lateral tunnel; ECC, extracardiac conduit; NR, not reported; HLHS, hypoplastic left heart syndrome; N/A, not applicable; AP, atriopulmonary; ICU, intensive care unit.

that dilation of the pathway (through enlarged conduits or dilated intra-atrial tunnels) results in substantial energy loss.¹⁹ Contrary to the above evidence, Bove and colleagues,²⁶ in their assessment of the fluid dynamics in various Fontan configurations, reported that the lowest energy loss was noted in LT Fontan, particularly following hemi-Fontan construction. They theorized that this may be due to the ease of caval offset and flaring of the cavopulmonary anastomosis, avoiding caval flow clashing and turbulence.

Although interesting, these fluid dynamic studies have not been able to profoundly influence our Fontan surgery and at this stage, do not seem to help us identify the superiority of one of these techniques.

SIZE OF FONTAN PATHWAY

One potential disadvantage of the ECC is its lack of capacity for growth. Any long-term outcomes may be affected by the size of conduit used. Early studies noted that IVC diameter at 2 to 4 years is approximately 80% of adult size, with significant variability among patients.²⁷ Several studies that examined ideal conduit size in relation to long-term outcomes and exercise capacity concluded that conduit oversizing may be more detrimental to outcomes owing to flow stagnation, energy loss, and the increased risk of thromboembolic events.^{28,29} In an exercise study analysis, Lee and colleagues³⁰ recently identified that the ideal conduit size is 12.5 mm/m² (16 to 18 mm on average) and that larger conduit sizes are worse. Others have claimed that Fontan-associated liver disease may be higher with ECC, especially with smaller conduits.³⁰

EARLY OUTCOMES

There is a considerable variation in mortality following the 2 techniques. Most would consider the early outcomes of these techniques equivalent, but 2 articles reported significant differences. Weixler and colleagues¹ described their experience with >800 patients from the Boston group between 2000 and 2017, with the majority undergoing a LT Fontan (n = 638). They reported a higher rate of early mortality following ECC Fontan compared with their LT cohort (3.1% vs 0.5%; $P < .01$), despite the fact that patients who underwent LT seemed to have more adverse risk factors. Patients who underwent LT had a higher incidence of right ventricular dominance and hypoplastic left heart syndrome (HLHS), and higher pulmonary artery pressure and pulmonary vascular resistance. A 2012 analysis of 2747 Fontan procedures performed in 68 centers between 2000 and 2009 recorded in the Society of Thoracic Surgeons (STS) database showed higher mortality in patients who underwent ECC Fontan (2.1% vs 0.9%; $P = .02$).⁸ This is in contrast to a recent large single-center study from Philadelphia of 773 patients from 1992 to 2009 that reported a higher overall operative mortality of 3.5%, with no difference between ECC and LT.³ In addition, the Australia and New Zealand Fontan Registry reported a low early mortality rate of 1.3% for ECC across a similar era (1997-2010) and up to 2.0% operative mortality for the LT technique ($P = .8$).^{6,31}

Early morbidity following Fontan operations can present as prolonged pleural effusions, hospital stay, and early Fontan takedown. The Boston group recently noted that despite longer cross-clamp times ($P = .01$) and bypass times ($P < .001$) in the LT cohort, there was no difference in early postoperative morbidity between the 2 techniques, other than

TABLE 2. Late outcomes of contemporary techniques

Authors	Dates	n	Follow-up, y (median)	Survival, %	Comparison	Comment
Weixler et al, 2020 ¹	2000-2017	Total: 801 LT: 638 ECC: 183	4.8	5 y: LT, 96.4 vs ECC, 92.0 10 y: LT, 94.5 vs ECC, 91.4 20 y: LT, 93.9 vs ECC, 90.8	Propensity-score matched: age, fenestration, heterotaxy, ventricular dominance, and concomitant procedures	Improved survival in LT group (HR, 2.7; $P < .05$). Better freedom from failure in LT group ($P = .02$). Pleural effusions more common in ECC group ($P = .01$).
Deshaies et al, 2019 ¹⁵	1974-2012	Total: 522 LT: 218 ECC: 192	11.6	5 y: 98.3 10 y: 94.7 20 y: 78.9	Demographics comparison between LT and ECC unavailable. Multivariable analysis of thromboembolic risk performed between LT and ECC (including arrhythmias and thromboprophylaxis).	Includes AP Fontan. No difference reported in survival or arrhythmias. Lower rate of thromboembolic events in ECC group compared with LT (HR, 0.34; 95% CI, 0.13-0.91).
Mery et al, 2019 ²	1995-2016	Total: 610 LT: 147 ECC: 463	6.8	5 y: 97 10 y: 94 15 y: 92	Demographics unavailable for Fontan-type comparison. Fontan type not considered a risk factor.	No difference reported in survival or failure. Higher incidence of arrhythmia in the LT group compared with the ECC group ($P = .002$).
Downing et al, 2017 ³	1992-2009	Total: 773 LT: 409 ECC: 364	9	10 y: 90 (95% CI, 88-92) 20 y: 74 (95% CI, 67-80)	Demographics unavailable for Fontan-type comparison. Fontan type included in multivariable risk factor analysis.	Survival with intact Fontan circulation. Fontan type did not impact late outcomes (survival or failure).
Iyengar et al, 2014 ⁶	1975-2010	Total: 1006 LT: 271 ECC: 532	NR	15 y: 93 (95% CI, 90-95) 20 y: 90 (95% CI, 86-93) 25 y: 83 (95% CI, 75-89)	Multivariable models accounting for common risk factors (including ventricular dominance and heterotaxy) performed to compare LT and ECC patients.	No difference reported in survival between LT and ECC. Higher rate of SVT in LT (HR, 3.1; $P = .019$). No reported difference in failure or other adverse events.
Nakano et al, 2015 ⁵	1994-2014	Total: 500 LT: 0 ECC: 500	7	15 y: 93	Only ECC	Only ECC in the study.
Ono et al, 2016 ⁴	1994-2015	Total: 434 LT: 50 ECC: 384	6.6	15 y: 92.3	ECC cohort had more right ventricular dominance, HLHS, heterotaxy, and fewer concomitant procedures and fenestrations at time of Fontan.	No reported difference in survival. Predicted % of peak VO_2 higher in ECC group ($P < .001$).

(Continued)

TABLE 2. Continued

Authors	Dates	n	Follow-up, y (median)	Survival, %	Comparison	Comment
Balaji et al, 2014 ¹⁶	1998-2008	Total: 1271 LT: 602 ECC: 669	9.2	NR	Univariable and multivariable analysis performed for ECC and LT comparison. ECC had more left ventricular dominance and older age at Fontan on univariable analysis.	Higher rate of early postoperative bradyarrhythmia in ECC group ($P < .001$). No difference at late follow-up between LT and ECC.
Robbers-Visser et al, 2010 ⁹	1988-2008	Total: 209 LT: 102 ECC: 107	4.3	Freedom from failure (mortality and failure) 3 y: LT, 85 vs ECC, 88 6 y: LT, 83 vs ECC, 79	Preoperative risk factors accounted for in multivariable analysis. HLHS and tricuspid atresia more common in ECC group.	No difference reported in survival/freedom from failure ($P = .93$). Higher rate of arrhythmia in the LT group on log-rank analysis ($P = .022$).
Hirsch et al, 2008 ¹⁴	1992-2007	Total: 636 LT: 585 ECC: 51	4	5 y: 95 10 y: 93 14 y: 91	ECC was performed in patients with anatomy not amenable to LT technique. Group comparison is inaccurate due to selection bias.	Unable to compare difference due to patient selection bias.
Mitchell et al, 2006 ¹⁷	1992-1999	Total: 322 LT: 262 ECC: 48	8.3	Freedom from death and transplantation 5 y: 94.9 (95% CI, 91.6-97.0) 8 y: 93.9 (95% CI, 90.1-96.2)	Comparison of demographics between LT and ECC unavailable. Univariable and multivariable analyses performed. No difference in preoperative or operative variables between group outcomes.	No difference reported in survival/freedom from failure or hospitalization ($P = .062$).

LT, Lateral tunnel; ECC, extracardiac conduit; HR, hazard ratio; AP, atriopulmonary; CI, confidence interval; SVT, supraventricular tachycardia; HLHS, hypoplastic left heart syndrome; VO₂, oxygen consumption; NR, not reported.

prolonged pleural effusions (3.9% for LT vs 8.6% for ECC; $P < .01$).¹ Similarly, the Australia and New Zealand cohort reported no difference between LT and ECC in early failure (6% vs 4%; $P = .2$), prolonged effusions (6% vs 8%; $P = .2$), or a composite of early adverse outcomes, including length of stay (17% vs 13%; $P = .7$).⁶ However, the STS database review, which accounted for patient characteristics, procedural factors, and center volume, found worse early outcomes on multivariable analysis in the ECC group with more takedown/revisions (0.7% for LT vs 1.7% for ECC; $P = .03$), increased length of stay (9 days vs 10 days; $P < .001$), and postoperative complications (32% vs 45%).⁸

LONG-TERM SURVIVAL, FREEDOM FROM HEART TRANSPLANTATION, AND FONTAN FAILURE

There have been several large publications of various Fontan cohort outcomes detailing the differences in outcomes between LT and ECC (Table 2). The Australia and New Zealand Fontan Registry reviewed the long-term outcomes of 271 LT and 532 ECC Fontan patients, which demonstrated relatively similar survival and freedom from failure in the 2 groups.³² Similarly, Downing and colleagues³ from Philadelphia noted that the type of Fontan did not predict early or late survival, freedom from transplantation, or Fontan failure.

The recent publication of Fontan outcomes from the Boston Children's Hospital has reopened this conversation. The long-term survival was better in the LT group compared with the ECC group (hazard ratio [HR], 2.7; $P < .05$), in contrast to most of the published data worldwide, which consistently demonstrates no difference between techniques.^{1,3,32,33} This difference in outcomes in the Boston group is also noted in their freedom from Fontan failure ($P = .02$) and their propensity score analysis; however, their propensity score analysis, which included heterotaxy syndromes and concomitant procedures, failed to show a significant difference in mortality at a 10-year follow-up.

Arrhythmias

Sinus node dysfunction and chronotropic incompetence have been shown to be common early and late after Fontan surgery and to impair exercise tolerance.^{6,32,34} Recently, chronotropic incompetence and decline in peak exercise capacity have been shown to predict mortality and the need for hospitalization and transplantation.³⁵ Avoidance of an atrial incision and suture lines is thought to reduce the rate of atrial arrhythmias, with their prevalence increasing with time. The exclusion of the atrium in the ECC Fontan also theoretically spares the atrial wall from distension and subsequent risk of arrhythmia. This theory is supported by Robbers-Visser and colleagues,⁹ who reported higher arrhythmic burden in their LT cohort compared with their ECC cohort ($n = 277$; freedom from arrhythmia, 83% vs 92% at 6 years; $P = .02$).⁹ In addition, a recent study from Texas of 610 patients reported higher rates of arrhythmia in the LT group ($P = .002$).² However, a large review by Balaji and colleagues of 1271 patients across 14 centers in 2014 reported no difference in the rate of late tachyarrhythmias between LT and ECC patients.¹⁶ Weixler and colleagues' recent comparison also noted no differences in pacemaker implantation, tachyarrhythmia (4.2% for LT vs 2.5% for ECC; $P = .3$) or sinus node dysfunction (4.9 for LT vs 1.8 for ECC; $P = .09$) between their 2 groups.¹

Clearly, the incidence of reported arrhythmias will differ with varying lengths of follow-up.^{36,37} Currently, there is no consensus on which technique results in higher arrhythmic burden.³⁸ As the contemporary Fontan patients age and their arrhythmic burden increases, the different management options will become more important. As Marshall and colleagues³⁹ reported in their recent review of Fontan arrhythmia management, lateral tunnel patients maintain transvenous atrial access if atrial pacing is required.

THROMBOEMBOLIC EVENTS

Thromboembolic events contribute to significant morbidity following the Fontan operation.⁴⁰ Until recently, there has been no consistently reported difference in thrombotic events between LT and ECC^{1,9,41}; however, in a recent

report by Deshaies and colleagues,¹⁵ a multivariable time-adjusted analysis of 522 patients across 12 institutions showed that ECC is associated with a lower incidence of thromboembolic events with a similar arrhythmic burden.¹⁵ The Boston group reported a large discrepancy in postoperative warfarin use between ECC and LT patients (33% vs 4.7%), which the authors attributed to pre-Fontan thromboembolic events.¹ Although no difference in overall post-Fontan thromboembolic events between the groups, the majority of early ECC deaths were attributed to thromboembolic complications. It is difficult to elucidate whether this is due the prosthetic material used in ECC or a preexisting tendency of these patients toward thrombosis. Neither pre-Fontan thrombosis nor warfarin use was included in the propensity score analyses, and differences that might have impacted their overall outcomes likely remained.

OTHER CONSIDERATIONS

The heterogeneity of patients undergoing the Fontan operations makes comparison of studies difficult. Heterotaxy syndromes and venous anomalies are common in the single-ventricle population, and there seems to be a tendency to favor ECC in this setting.⁸ In patients with interrupted IVC, the Kawashima procedure can be useful; however, there is inevitable formation of pulmonary arteriovenous malformations. A technique with a Y-graft ECC has been theorized to equally distribute hepatic flow (and thereby hepatic factor) to both lungs.⁴² Another variation that can be useful in complex rerouting of the systemic venous flow is the intra/extracardiac Fontan, originally described by Vargas and colleagues.⁴³

DISCUSSION

The surgical technique of the Fontan procedure has evolved since its conception but we are still uncertain whether the lateral tunnel and the extracardiac conduit, the most widely used techniques, are equivalent. Both were studied with a plethora of fluid dynamics studies to determine the ideal Fontan pathway. The early work by de Leval and others helped identify large pitfalls in constructing the Fontan pathways incorporating the entire atrium and highlighted the importance of creating the most streamlined pathway.^{19,24} However, as Tang and colleagues²⁵ demonstrated, there is significant variability within each of these techniques. These studies have not yet helped us to identify the relative superiority of LT and ECC.

There was hope that patients operated with the extracardiac conduit would have less arrhythmia, and globally, there seems to have been a large shift toward performing ECC over LT.^{7,15,32} However, after almost 3 decades, there is little evidence of the superiority of one technique over the other in terms of long-term outcomes.

Importantly, there is recent evidence of worse early outcomes after ECC compared to the LT technique.¹ Published

rates of operative mortality vary substantially, particularly for the ECC technique.^{1,3,6} This variability might be the reflection of the technical challenges of performing this operation accurately. The earlier fluid dynamics studies have clearly demonstrated that small changes in anastomotic angles and offset have profound effects on flow. In addition, the anastomosis of an extracardiac conduit to the pulmonary arteries carries the potential for significant distortion of the PAs, particularly at a young age. Technical issues such as these may present as early or late adverse outcomes and have recently been demonstrated to contribute substantially to patient mortality.⁴⁴

Despite the simple design of the extracardiac conduit, its realization and accurate reproducibility is more complicated. It should be noted that groups who demonstrated better results with the extracardiac conduit, such as the Australia and New Zealand group, appear to proceed with the Fontan operation at a later age (mean, 4.7 years) compared with many of the larger North American institutions.^{1,3,6} Performing the ECC technique in larger patients may reduce IVC–conduit mismatch and minimize PA distortion.

There is significant heterogeneity in the patients requiring the Fontan operation and it is possible that one of these techniques may be better suited for specific patients. For example, the pulmonary arterial and systemic venous anomalies and apicocaval juxtaposition frequently encountered in heterotaxy syndromes may be better served by one of the techniques, depending on the anatomy. In addition, patients with HLHS, who have been noted to develop arteriovenous malformations more frequently and thus may require earlier Fontan completion, may be better served by LT Fontan. It should be noted that in most series describing late outcomes, the reason to choose one or the other technique were not specified, other than institution or surgeon preference.

CONCLUSIONS

Whether LT or ECC is the superior technique remains controversial. Reports of worse early adverse outcomes after ECC may reflect the technical complexities of accurately performing this technique. As institutions and surgeons tend to favor one technique over the other, comparison of outcomes has become difficult. Because of this equipoise, one may wonder whether further prospective research will be able to identify which technique should be used in specific circumstances.

Conflict of Interest Statement

Dr d'Udekem is a consultant for Actelion.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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